TABLE 134-9 INTERNATIONAL PROGNOSTIC INDEX FOR NON-HODGKIN'S LYMPHOMA

Five clinical risk factors:	
Age ≥60 years	
Serum lactate dehydrogenase levels elevated	
Performance status ≥2 (ECOG) or ≤70 (Karnofsky)	
Ann Arbor stage III or IV	
>1 site of extranodal involvement	
Patients are assigned a number for each risk factor they have	
Patients are grouped differently based on the type of lymphoma	
For diffuse large B-cell lymphoma:	
0, 1 factor = low risk:	35% of cases; 5-year survival, 73%
2 factors = low-intermediate risk:	27% of cases; 5-year survival, 51%
3 factors = high-intermediate risk:	22% of cases; 5-year survival, 43%
4, 5 factors = high risk:	16% of cases; 5-year survival, 26%
For diffuse large B-cell lymphoma treated with R-CHOP:	
0 factor = very good:	10% of cases; 5-year survival, 94%
1, 2 factors = good:	45% of cases; 5-year survival, 79%
3, 4, 5 factors = poor:	45% of cases; 5-year survival, 55%

Abbreviations: ECOG, Eastern Cooperative Oncology Group; R-CHOP, rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone.

gallium scans are much more useful in aggressive subtypes such as diffuse large B-cell lymphoma than in more indolent subtypes such as follicular lymphoma or small lymphocytic lymphoma. Although the IPI does divide patients with follicular lymphoma into subsets with distinct prognoses, the distribution of such patients is skewed toward lower-risk categories. A follicular lymphoma–specific IPI (FLIPI) has been proposed that replaces performance status with hemoglobin level (<120 g/L [<12 g/dL]) and number of extranodal sites with number of nodal sites (more than four). Low risk (zero or one factor) was assigned to 36% of patients, intermediate risk (two factors) to 37%, and poor risk (more than two factors) to 27% of patients.

CLINICAL FEATURES, TREATMENT, AND PROGNOSIS OF SPECIFIC LYMPHOID MALIGNANCIES

PRECURSOR CELL B-CELL NEOPLASMS

Precursor B-Cell Lymphoblastic Leukemia/Lymphoma The most common cancer in childhood is B-cell ALL. Although this disorder can also present as a lymphoma in either adults or children, presentation as lymphoma is rare.

The malignant cells in patients with precursor B-cell lymphoblastic leukemia are most commonly of pre–B cell origin. Patients typically present with signs of bone marrow failure such as pallor, fatigue, bleeding, fever, and infection related to peripheral blood cytopenias. Peripheral blood counts regularly show anemia and thrombocytopenia but might show leukopenia, a normal leukocyte count, or leukocytosis based largely on the number of circulating malignant cells (Fig. 134-5). Extramedullary sites of disease are frequently involved in patients who present with leukemia, including lymphadenopathy, hepato- or splenomegaly, CNS disease, testicular enlargement, and/or cutaneous infiltration.

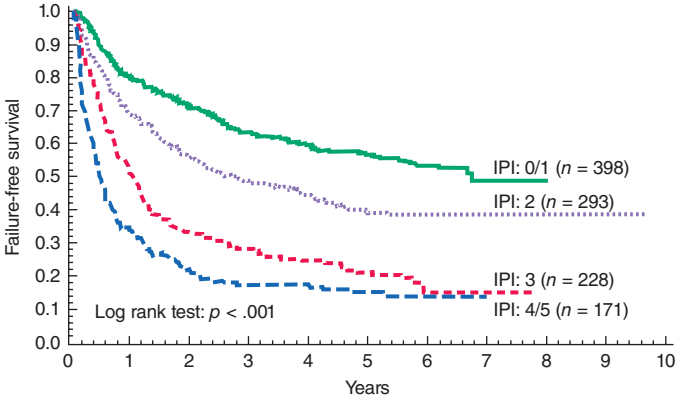

FIGURE 134-4 Relationship of International Prognostic Index (IPI) to survival. Kaplan-Meier survival curves for 1300 patients with various kinds of lymphoma stratified according to the IPI.

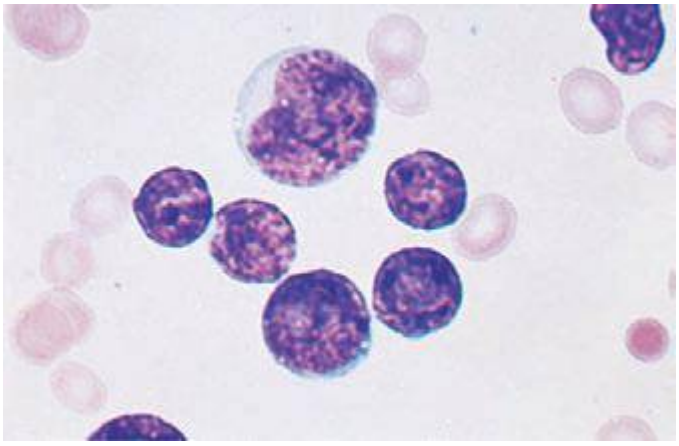

FIGURE 134-5 Acute lymphoblastic leukemia. The cells are heterogeneous in size and have round or convoluted nuclei, high nuclear/cytoplasmic ratio, and absence of cytoplasmic granules.

The diagnosis is usually made by bone marrow biopsy, which shows infiltration by malignant lymphoblasts. Demonstration of a pre–B cell immunophenotype (Fig. 134-2) and, often, characteristic cytogenetic abnormalities (Table 134-6) confirm the diagnosis. An adverse prognosis in patients with precursor B-cell ALL is predicted by a very high white cell count, the presence of symptomatic CNS disease, and unfavorable cytogenetic abnormalities. For example, t(9;22), frequently found in adults with B-cell ALL, has been associated with a very poor outlook. The bcr/abl kinase inhibitors have improved the prognosis.

TREATMENT PRECURSOR B-CELL LYMPHOBLASTIC LEUKEMIA

The treatment of patients with precursor B-cell ALL involves remission induction with combination chemotherapy, a consolidation phase that includes administration of high-dose systemic therapy and treatment to eliminate disease in the CNS, and a period of continuing therapy to prevent relapse and effect cure. The overall cure rate in children is 90%, whereas ~50% of adults are long-term disease-free survivors. This reflects the high proportion of adverse cytogenetic abnormalities seen in adults with precursor B-cell ALL.

Precursor B-cell lymphoblastic lymphoma is a rare presentation of precursor B-cell lymphoblastic malignancy. These patients often have a rapid transformation to leukemia and should be treated as though they had presented with leukemia. The few patients who present with the disease confined to lymph nodes have a high cure rate.

MATURE (PERIPHERAL) B-CELL NEOPLASMS

B-Cell Chronic Lymphoid Leukemia/Small Lymphocytic Lymphoma B-cell CLL/small lymphocytic lymphoma represents the most common lymphoid leukemia, and when presenting as a lymphoma, it accounts for ~7% of non-Hodgkin's lymphomas. Presentation can be as either leukemia or lymphoma. The major clinical characteristics of B-cell CLL/small lymphocytic lymphoma are presented in Table 134-10.

The diagnosis of typical B-cell CLL is made when an increased number of circulating lymphocytes (i.e., >4 × 10^9/L and usually